Diabetes Mellitus in a Patient with Aarskog Syndrome on Growth Hormone Treatment

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Introduction

It is well known that growth hormone (GH) has an anti-insulin effect(1). However, the number of reported patients with secondary diabetes mellitus is limited(2).

Case Report

We report a case of diabetes mellitus in a 12-year-old boy with Aarskog syndrome. The diagnosis was made at the age of 10 years because of his peculiar facies (broad nasal bridge, hypertelorism, widow's peak), typical features (interdigital webbing, brachydactyly, shawl scrotum, cryptorchism), and short stature. His gestational age was 37 weeks and his birth weight was 3,125 g, with normal cephalic delivery.

He had been diagnosed since birth as having an atrial septal defect, with chronic thrombocytopenic purpura associated with positive lupus anticoagulant antibody.

His height was 133 cm (−2.2 SD) and he
weighed 30 kg (−1.9 SD) at the age of 11 years and 10 months. His height velocity was 3.3 cm/year, his skeletal maturation was consistent with his age (bone age 13 years by the Greulich & Pyle method) and pubertal development was Tanner stage II.

He had peak GH values of 0.62, 10.1, and 5.70 ng/mL after arginine, insulin and L-dopa stimulation respectively. We started daily subcutaneous injections of GH at a dose of 0.5 IU/kg/week in November 1994. Glycosuria was noticed one month later and hyperglycemia was evident at 153 mg/dL in Jan. 1995. Though GH therapy was stopped immediately, his glycosuria persisted even 4 months after cessation of GH therapy. An oral glucose tolerance test showed a diabetic pattern at that time. He was then put on diet therapy (1,800 cal/day), which resulted in only slight hyperglycemia and improved glucose tolerance.

As his growth velocity was very low at 1 cm/year, we decided to start GH treatment again very carefully in June 1995. At present, one month after initiation of GH, his urinary glucose excretion is 3–5 g/day, and fasting blood glucose remains normal.

### Discussion

Among patients with Aarskog syndrome, several cases have been reported to have GH deficiency (3). However, in only one such case was GH therapy prescribed (4), and detailed information was not available.

It seems obvious that initiation of GH therapy induced glucose intolerance in this case. However, limited capacity of insulin secretion was suggested by the repeated oral
Diabetes Patient with Aarskog Syndrome on GH Glucose tolerance tests (OGTTs), while administration of GH therapy may have coincided with insulin resistance. With regard to risk factors for diabetes, the family history was negative, and he was not obese. Aarskog syndrome has not yet been known to predispose to diabetes. Although the islet cell antibody test was negative, an autoimmune mechanism may be involved in this case, considering the coexisting thrombocytopenic purpura associated with positive lupus anticoagulant antibody and PAIgG.

References


